Images In Medicine: Primary Tracheobronchial Amyloidosis

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Abstract

We present a case of a non-smoker 23-year-old man who presented complaining of progressive shortness of breath and productive cough for 2 months. A diagnosis of tracheobronchial amyloidosis was made.

CASE REPORT

A non-smoker 23-year-old man has presented complaining of progressive shortness of breath and productive cough for 2 months. Physical examination revealed mild parasternal sibling, and past medical records were unremarkable. Initial chest computed tomography (CT) "hands up" scout demonstrated a tortuous trachea, without paratracheal stripes blurring (Figure 1).

Figure 1

Figure 1. Chest computed tomography (CT) scouth demonstrating tracheal tortuous enlargement, without – however – blurring paratracheal stripes.



A chest Computed Tomography (CT) scans corroborated

such finding, and indicated the presence of diffuse circumferential tracheal thickening, with multiple foci of calcifications (Figure 2), raising the diagnostic suspicion of amyloidosis.

Figure 2

Figure 2. Chest computed tomography (CT) indicating diffuse circumferential tracheal thickening, associated with multiple calcified foci, which is very useful for differential diagnosis.



3D-volume-rendering (3DVR) CT tool (Figure 3) and the

performance of virtual bronchoscopy (Figure 4) were used to confirm and better assess anatomical abnormalities.

Figure 3

Figure 3. 3D-volume-rendering (3DVR) CT tool, corroborating imaging features and providing better anatomical detailing.



Fiberoptical bronchoscopy samples were positively stained with Congo red at histochemical analysis, corroborating the diagnosis. Further screening investigation for other organ involvement was negative, conferring the primary character for tracheobronchial amyloidosis.

Figure 4

Figure 4. Virtual bronchoscopy CT tool performance providing intraluminal appearance of tracheobronchial amyloidosis, with multiple irregular affected areas.



Tracheobronchial amyloidosis is a rare disease, in which CT currently plays a diagnostic role, once clinical findings are frequently non-specific. 1,2 The characteristic circumferential involvement, without sparring tracheal muscular components (posterior wall) is very useful for differential diagnosis establishment with osteocondroplastic tracheobronchopathy, polychondritis recidivans and Mounier-Kuhn disease, which typically affects only cartilaginous structures. While videoguided stenotic resection is usually the therapy of choice, interventional decision should always be held on an individual basis. 1

References

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